TOPIC 17 – Congenital heart diseases

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0126

One-stage percutaneous Melody pulmonary valve implantation: a single-center experience in 21 consecutive patients

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Purpose: Right ventricular outflow tract (RVOT) reconstruction in infancy or childhood leads to lifetime iterative surgery for pulmonary stenosis or regurgitation. Transcatheter pulmonary valve implantation (TPVI) was developed with the aim to reduce the number of reoperations and operative-related morbidity.

Methods and Results: From 12/2008 to 03/2010, 21 patients were referred to Marie-Lannelongue Hospital for TPVI. Mean age was 24 yrs (11.5-55 yrs), mean weight was 60 kg (20-85 kg). Most of them had a Tetralogy of Fallot variant (n=12) and 70% had 3 or more previous surgery. RVOT stenosis was predominant in 90% of patients. RVOT reconstruction techniques were: valved conduit (n=12), non-valved conduit (n=1), homograft (n=6), RVOT patch (n=2). RVOT balloon inflation test was performed only if cardiac CT scan showed coronary arteries located next to the RVOT. Two patients were excluded, 1 because an aorto-pulmonary fistula after Ross operation, and 1 with previous arterial switch because of compression of the reimplanted left coronary artery at RVOT balloon inflation test. Initial stenting and TPVI were performed during the same procedure in 18/19. There was immediate significant decrease in right ventricular systolic pressure (mean 88,6 to 52,2 mmHg), mean RVOT gradient (mean 43,7 to 22 mmHg) and pulmonary regurgitation (grade 2 or more before, none after). One patient with a 16mm homograft had significant residual stenosis (mean RVOT gradient 45 mmHg) after Melody implantation. All patients were discharged within 2 days. At a mean follow-up of 9.3 months (1 to 24 months) all patients are alive and well and neither Melody valve dysfunction nor stent fracture or migration was noticed.

Conclusion: Percutaneous pulmonary valve implantation is a safe alternative to surgery. Our experience suggests that patients with RVOT conduit<18 mm and those with previous arterial switch operation should be referred to surgery without considering transcatheter procedure.

0127

Characteristics and long-term outcome of non-immune isolated atrioventricular block diagnosed in utero or early childhood: a multicenter study

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Background: The natural history of isolated, non-immune congenital or childhood atrioventricular (AV) block is poorly known.

Methods and Results: We retrospectively studied 141 children with isolated, non-immune AV block diagnosed *in utero*, or up to 15 years of age, at 13 French medical centers, between 1980 and 2009. Patients with structural heart disease or maternal antibodies were excluded. AV block was asymptomatic in 119 (84.4%) and complete in 100 (70.9%) patients. Incomplete AV block progressed to complete in 29 (70.7%) patients with incomplete block over 2.8±3.4 years (1-155 months). Narrow QRS complexes were present in 18 of 26 patients (69.2%) with congenital, and 106 of 115 (92.2%) with childhood AV block. Pacemakers were implanted in 112 children (79.4%), during the first year of life in 18 (16.1%) and before 10 years of age in 90 (80.4%). The mean delay between diagnosis of AV block and pacemaker implants was 2.6±3.9 years (0-300 months). The pacing indication was prophylactic in 70 children (62.5%). During a median follow-up of 11.6±6.7 years (1-32 years), no patient died or developed dilated cardiomyopathy. The long-term follow-up was uncomplicated in 127 children (90.1%).

Conclusions: In this large multicenter study, the long-term outcome of isolated, non-immune congenital or childhood AV block was favorable, regardless of the patient's age at the time of diagnosis. No patient died or developed dilated cardiomyopathy, and pacemaker-related complications were few. The progression of incomplete to complete AV block in nearly 70% of patients suggests a postnatal degeneration of the specialized conduction system.

0435

Tetralogy of Fallot: impact of pulmonary valve replacement on left ventricular function

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Pulmonary valve regurgitation is the most common mid- to long-term complication after complete repair of Tetralogy of Fallot (ToF). The benefit of Pulmonary Valve Replacement (PVR) on right ventricular volumes is well established. However, impact on left ventricular function is still debated. We sought to determine the evolution of left ventricular function after PVR. In this abstract we report the preliminary results of this monocentric prospective ongoing study.

Methods: Patients with a history of complete repair of ToF and requiring a PVR are prospectively enrolled. They all undergo a cardiac MRI (CMR) performed before and after the PVR at 1.5 Tesla (Intera®; Philips Medical Systems, Best, The Netherlands). The CMR protocol includes cine steady-state free precession sequences in short axis planes (slice thickness=5mm; space between slices=5mm) to assess the ventricular volumes. Left ventricular end-diastolic (EDV), end-systolic volumes (ESV) and ejection fraction (EF) are calculated from manual contouring of the left ventricular chamber. A Wilcoxon test is performed to compare EF before and after the PVR.

Results: 13 patients (mean age 32 +/- 15 years) has been included between July 2009 and January 2011. EDV and ESV were respectively 66.3±21.7 and 34.8±15.4mL/m² before the PVR and 72.3±13.0 and 32.8±10.6 mL/m² after, leading to a significant increase in EF of 7.2±8.6% (48.1±10.1% before PVR and 55.3±9% after PVR, p=0.01).

Conclusions: The preliminary results of this ongoing study show a significant improvement in left ventricular function after PVR. Further investigations are needed to understand the underlying mechanisms of this evolution.

0320

Multi facetted circulation monitoring during the first 72 hours after switch operation

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Background: Monitoring of circulation and cardiac output is a priority in the paediatric cardiac ICU, especially in cases of post surgical heart failure. For this purpose, clinical, biological and technical alternatives are available. But which of them is the most reliable?

Methods: We examined 30 consecutive newborns undergoing switch operation by clinical, biological and technical means for cardiac and circulatory functions. The clinical parameters were measured every 6 hours: heart rate, mean blood pressure and diuresis. As biological parameters served (every 6 hours): Troponine Ic, BNP, Renine, Lactate. Technical means for the evaluation of cardiac function and output were transthoracic echocardiography (GE, Vivid 7) every 12 hours and continuous bioimpedance measurement (Osypka, Aesculon).

Results: During the first 24 hours after switch operation for simple TGA, blood pressure fell, heart rate raised and diuresis slowed down significantly ($p<0.01$). BNP first rose (H6) then fell slowly ($p<0.01$), renine reacted inversely during the first 36 hours, and then behaved parallel to BNP. Troponine and lactates showed linear kinetics. Cardiac output fell to 50%. Bioimpedance and measurement of cardiac output by transthoracic echocardiography correlated well ($r=0,84$). Recovery set on at H24-36 and ameliorated significantly in all parameters ($p<0.01$). None of the patients showed clinical haemodynamic or respiratory complications.

Conclusions: diagnosis of the circulatory status of a newborn in the first 72 hours after switch operation can be refined by adding BNP and renine measurement in addition to the "classical parameters". Bioimpedance shows the same reliability as echocardiographical exams with the advantage of a continuous monitoring, thus assuring more security for the patient.

0322

Electrical velocimetry as a tool for cardiac output measuring in small infants with heart failure

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Objectives: We investigated if electrical velocimetry (EV), would be interchangeable with transthoracical echocardiographical Doppler (TTE) for the measurement of cardiac output (CO) in the pediatric intensive care unit (PICU) and if it was able to follow the post surgical trend of CO correctly with the advantage of continuous measurement.

Background: CO is an essential circulatory component to guarantee tissue and organ perfusion. Especially in the intensive care setting, acute heart failure is disastrous for the patient. As invasive CO measuring methods may cause complications in children, and TTE needs an expertise perhaps not available in a non cardiologic PICU, other non invasive alternatives become important.

Methods: Comparison of stroke volume (SV) measurements by EV and TTE was made in 24 newborns after switch operation (n=240 measurements). 3 subgroups of measurements were created according to the stadium in the course of post surgical recovery of cardiac output.

Results: In the Bland – Altman plot, acceptable bias (0.27 ml) and limits of agreement were found indicating that the two methods were interchangeable. Correlation was good with $r=0.86$, regression showed good predictability. Overall percentage error was acceptable with 25.8%. SV Medians (either by SV-EV or SV-ETT) in the three subgroups were different, and precision was clearly below 20% indicating the usefulness of both methods to document CO trends.

Conclusions: EV and TEE are interchangeable non invasive methods for estimating CO correctly. Continuous CO measurement by EV may help to detect or control heart failure in the PICU in time.

0324

Non invasive ventilation diminishes post extubational afterload rise in small infants with heart insufficiency after cardiac surgery

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Objectives: Extubation may be of risk in critically ill small infants after cardiac surgery, resulting in pulmonary, metabolic and circulatory imbalances. Non invasive post extubational ventilation (NIV) may be a respiratory support. The aim of this study was to investigate if there would also be a positive effect of NIV on circulation, using BNP as a parameter reflecting ventricular wall stress (cut off for heart insufficiency laid at 400 pg/ml).

Methods: 34 infants (median age 9 (3 – 150) days, median weight 3.6 (2.8 – 5.0) kg, BNP > 400 pg/ml) after cardiac surgery for coarctation, tetralogy of Fallot, pulmonary atresia, TGA, AVSD. They were weaned from ventilation by 30 minutes of CPAP + inspiratory aid (IA) with PEEP=4 cmH2O, IA=12 cmH2O, FiO2=0.4, then extubated under stable clinical and blood gas conditions. 15 of them at random received NIV (CPAP, PEEP=4 cmH2O, FiO2 0.4) after extubation. Respiratory, metabolic and clinical circulatory parameters as well as BNP were measured before, 1 and 6 hours after extubation.

Results: In both groups, no significant difference of the clinical parameters developed after extubation. In the untreated group BNP rose significantly from median 725 pg/ml before extubation to 952 and 1280 pg/ml at 1 and 6 hours after extubation, respectively ($p=0.003$). Whereas in the NIV group, there were no significant changes in BNP (867, 818 and 1030 pg/ml before, 1 and 6 hours after extubation, respectively, $p=0.423$).

Conclusion: Extubation in infants suffering from heart insufficiency after cardiac surgery raises ventricular afterload. This subliminal heart insufficiency may clinically gain in consequence in the presence of respiratory or metabolic problems. Non invasive ventilation can prevent the afterload rise and should therefore be considered as a potential circulatory support in the post extubational course.

0323

Innovation: Catheter Interventions in Congenital Heart Disease without Catheterization Laboratory Equipment, The Chain of Hope Experience in Rwanda

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Introduction: Thousands of children live in developing countries with untreated but correctable congenital heart disease and most of them will die due to the lack of suitable medical and surgical facilities.We report the feasibility and safety of cardiac catheterization in a developing country without access to a regular cardiac catheterization laboratory.The equipment used for imaging consisted of a mono-plan conventional C-Arm X-Ray system and a portable ultrasound machine.

Method: During a week,for 4 consecutive years (2007-2010),a team of 2 paediatric cardiologists,1 anesthetist,2 nurses and 1 technician from the Chain of Hope-Belgium performed cardiac catheterisation at King Faiçal Hospital-Kigali, to treat 47 patients (45 children - 2 adults).This team was working with local personnel (doctors,nurses and technicians) to facilitate the transfer of expertise.At the same moment and in the same hospital,an australian surgical team was performing open heart surgery,providing a surgical back-up in case of catheterization complication.

Results: Treatment,using usual guide-wires and catheters for cardiovascular access,was successful in 45 out of the 47 patients,consisting in 29 patent ductus arteriosus occlusions with coils (n=6) or Amplatzer device (n=23);3

secundum ASD closure,12 pulmonary valve stenosis (PS) dilatation (3 critical PS,1 PS in a Fallot patient,2 PS in patient with ASD device closure),3 dilatation of ao coarctation.Two cases required surgery,1ASD closure after embolisation of an ASD device,1 PDA to large for device closure.No other complication was observed and most of the patients were discharged home the day after cardiac catheterization with good evolution on follow-up.

Conclusion: Cardiac catheterization can be performed safely and is highly effective in a country with limited resources.This mode of treatment is possible without the support of a sophisticated catheterization laboratory. Working with local staff allows the transfer of expertise.

0360

Exercise testing coupled with Doppler echocardiography: a simple, safe and effective method to assess the severity of coarctation of the aorta

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Peak systolic Doppler velocity at the aortic isthmus alone is not a good predictor of severity in patients with native or residual coarctation after surgical repair. Evaluating the need for surgery or percutaneous stenting is often difficult in these cases. A major argument in favour of a hemodynamically significant aortic isthmus narrowing is the presence of a diastolic gradient with characteristic “sawtooth” appearance of the Doppler pattern.

Objectives: to evaluate, in patients with suspected aortic isthmus narrowing but without any significant diastolic gradient at rest, if exercise could unmask diastolic gradient and thus reveal a hemodynamically significant coarctation.

Methods: Fourteen patients aged from 12 to 56 years underwent treadmill exercise testing coupled with Doppler echocardiography. Thirteen had previous coarctation repair 8 to 43 years before, one had aortic kinking with mild diffuse isthmic narrowing on CT-scan. MRI or CT-scan were performed in 11 patients, significant residual stenosis (narrowing >30%) was detected in 8/11. Exercise testing was maximal or juxtamaximal in all but 1. Doppler measurements were performed during exercise testing and 5 minutes after at the suprasternal notch, using a CW Doppler 2 MHz pedoff probe. The peak systolic and diastolic gradients through aortic isthmus were measured at rest and at the end of the exercise. A peak protodiastolic gradient > 17 mmHg was considered as significant, as described in previous studies.

Results: Mean peak systolic gradient increased from 29 to 65 mmHg with exercise (mean increase +142%, $p<0.001$). Among the eight patients without any diastolic gradient at rest, significant diastolic gradient appeared in 4. Among the 6 patients with non significant diastolic gradient at rest, the gradient remained non significant in 2 and became significant in 4.

Conclusion: Apparition of a significant isthmic Doppler diastolic gradient at exercise can reliably predict the hemodynamic significance of aortic restenosis after coarctation repair, as well as native narrowing of the aorta. This simple, safe and effective non-invasive method may be helpful to identify patients requiring surgery or percutaneous stenting. It could also be useful in the follow-up of patients with a coarctation operated on in infancy.